

## Atypical Dermoid Presentations-Histopathological Diagnosis and Surgical Management

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### Abstract

The commonest cysts of the orbit are dermoid and epidermoid which are lined by keratinizing squamous epithelium. Dermoid typically present during childhood, but can present at any age. It has been observed that those presenting later in life are often deeper in origin and more challenging in terms of management. Superficial dermoid most often present as an asymptomatic, mobile mass. Deeper tumors may cause diplopia and proptosis. They typically occur superotemporally in the orbit. Other orbital sites are much less common. Four cases with atypical dermoid presentation that consulted a tertiary eye care center were reported. The clinical and radiological features, surgical approach and histological findings are described. Out of the four cases, three were confined to the orbit while one presented as a conjunctival mass. None of them showed any bony invasion. In the orbit, one mass was found over the brow over the frontal bone while the other occurred inferomedially and medially, all unusual sites for dermoid. Excision biopsy of these lesions confirmed dermoid cyst on histopathology. These atypical dermoids although rare do occur.

**Keywords:** Orbit; Atypical; Dermoid cyst

### Introduction

Dermoid is a developmental choristoma lined with epithelium and filled with keratinized material. 10% of head and neck dermoid cyst are localized to the orbit. They constitute 24% of all orbital lid masses [1]. 60% are located in upper outer quadrant. Dermoid cysts originate from aberrant ectodermal tissue. These tumors contain normal-appearing tissue in an abnormal location. As two suture lines of the skull close during embryonic development, dermal or epidermal elements may be pinched off and form cysts, which are adjacent to the suture line.

Histologically, dermoids are composed of epidermal tissue together with one or more dermal adnexal structures and skin appendages, such as hair follicles, sebaceous glands, and sweat glands. The cystic component is lined by keratinizing epidermis and may be filled with keratin, hairs, and fatty material. If the cyst wall does not show any adnexal structures common to skin, then the diagnosis of epidermoid is warranted. While dermoids and epidermoids differ histologically, their presentation and management are identical [2].

Dermoid cyst typically presents during childhood, but can present at any age [3]. It has been observed that those presenting later in life are often deeper in origin and more challenging in terms of management. Superficial dermoid most often present as an asymptomatic, mobile mass. Deeper tumors may cause diplopia and proptosis. Orbital inflammation from leakage of cyst contents or cyst rupture following blunt trauma can also be a presenting sign. Anterior

dermoid are located either superolaterally around the frontozygomatic suture or superomedially around frontoethmoidal or frontolacrimal sutures. Deep dermoid are located near the sphenozygomatic or sphenoethmoidal suture.

CT scan is extremely valuable in establishing the diagnosis of dermoid tumors [4]. They characteristically appear cystic and arise in close relationship to bone.

As dermoid cyst arise from the sequestration of surface ectoderm by closure of the underlying suture lines of the bony orbit. Consequently, they can arise from virtually any suture site and present in a variety of ways depending upon the suture of origin. We thus present a case series of dermoid cysts which have atypical location of presentation, confirmed histopathologically and their surgical management.

### Case Series

#### Case 1

A 35 year old male presented with a firm painless swelling below the right inner canthus. According to him, the swelling was noticed at the time of birth and has increased to the present status. It was a well-defined painless firm swelling not fixed to the skin or underlying structure (Figure 1a). On examination visual acuity was 20/20, with normal anterior segment and normal ocular movements. CT scan of the orbit showed a well-defined soft tissue density lesion at infero-medial aspect of right orbit. No bony erosion was found.

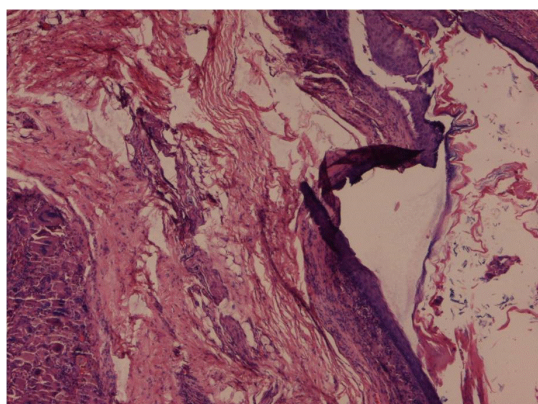


**Figure 1a:** Showing swelling below the right inner canthus.

Lesion was excised through medial orbitotomy via lynch incision with minimal scarring (Figure 1b). On histopathological examination there were findings consistent with dermoid cyst i.e. stratified squamous epithelium with dermal appendages (Figure 1c).



**Figure 1b:** Showing good post-operative cosmesis.



**Figure 1c:** Photomicrograph showing stratified squamous epithelium with dermal appendages (H&E, 100X).

## Case 2

A 16 year old girl presented with complaints of a growth present in the left brow region since childhood. According to the parents they noticed it just after birth and the size has increased since then. On examination it was a firm swelling 3 cm × 2.5 cm not attached to the skin or underlying structures. It was freely mobile over the bone, and skin over it was also free (Figure 2a). CT scan revealed well circumscribed non enhancing hypodense cystic lesion over the left frontal bone.



**Figure 2a:** Showing swelling at left brow region.

Excision biopsy via a lid crease incision was performed (Figure 2b). On gross examination, keratin and hair follicles can be appreciated (Figure 2c).



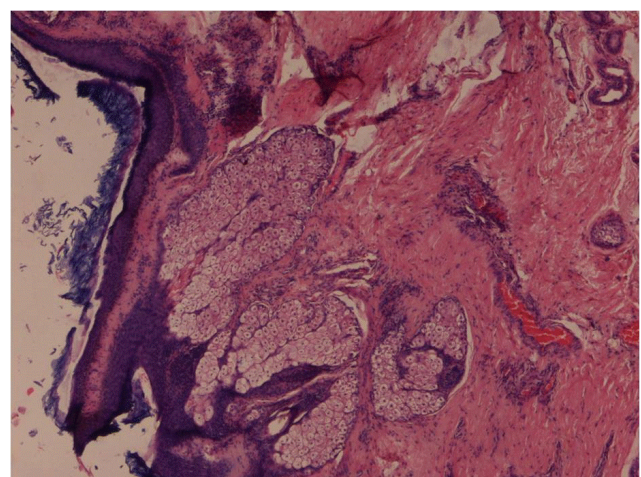
**Figure 2b:** Showing lid crease incision.

Histopathological examination of tissue section showed hyperkeratotic stratified squamous epithelium along with sebaceous glands, fibrofatty aggregates, hair follicle, with keratinous debris and foreign body inflammatory reaction, consistent with dermoid cyst (Figure 2d).





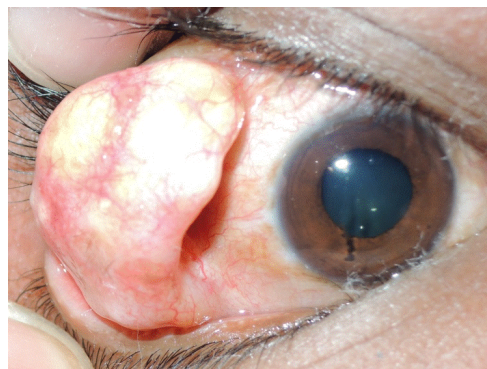
**Figure 2c:** Showing keratin and hair follicles on gross appearance.



**Figure 2d:** Photomicrograph showing hyperkeratotic stratified squamous epithelium with sebaceous gland, fibro-fatty tissue, hair follicle (H&E, 100X).

### Case 3

A 30 year old male presented with swelling in lateral canthus of right eye since birth. It was painless, multilobulated, firm swelling. There was gradual progressive increase in swelling for past 3-4 years. On examination a single pinkish yellow globular, multilobulated 15 mm × 10 mm was present in the lateral canthus in the interpalpebral area (Figure 3a). On slit lamp biomicroscopy hair follicles and hairs were visible on surface of swelling. MRI showed a well-defined altered signal intensity lesion at the outer canthus of right orbit which appeared hypointense in the centre and hyperintense in the periphery.

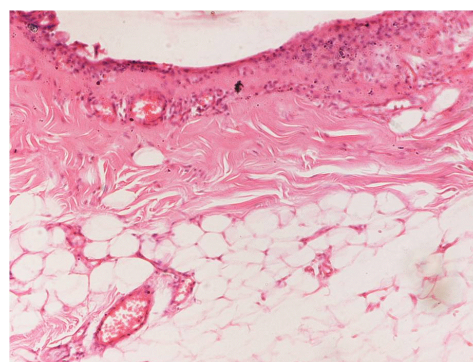


**Figure 3a:** Showing multilobulated swelling at lateral canthus.

Excision of the swelling was done via trans conjunctival route (Figure 3b). Histopathological examination revealed stratified squamous lined spaces in fibrocollagenous background, with sebaceous glands and abundant adipose tissue consistent with dermoid cyst (Figure 3c).



**Figure 3b:** Showing excision via transconjunctival route.



**Figure 3c:** Photomicrograph showing stratified squamous lined spaces in fibrocollagenous background, with sebaceous glands and abundant adipose tissue (H&E, 100X).

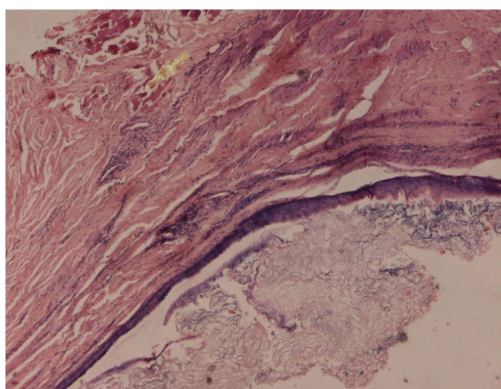
#### Case 4



**Figure 4a:** Showing mass at medial canthus area.



**Figure 4b:** Showing lynch incision for excisional biopsy.



**Figure 4c:** Photomicrograph Hyperkeratotic stratified squamous epithelium with skin appendages (H&E, 100X).

A 12 yrs old male presented with a painless swelling near the medial canthus since childhood. On examination it was a mobile, firm swelling of 8 mm × 9 mm (Figure 4a). The ocular movements were normal. CT revealed small hypodense moderately enhancing lesion

near the medial canthus of right orbit. There was no bony invasion seen.

Excision of the mass was performed by giving a Lynch incision medially and the mass was sent for histopathological examination which confirmed the diagnosis of dermoid cyst (Figures 4b and 4c).

#### Discussion

Some atypical presentations of dermoid in the past have drawn our attention. One is the case of an orbital floor dermoid. 18-year-old female patient presented with progressive, painless swelling in the lower eyelid associated with mild proptosis of three months duration. Lesion was excised and diagnosed as dermoid cyst on histopathological examination [5].

Another unusual presentation of orbital dermoid cyst causing superior oblique muscle palsy in a child was reported. A dermoid cyst at the region of trochlea was suspected as the cause of superior oblique muscle palsy in this case [6].

In another study, five cases of atypical orbital cystic choristomas were reported, Three of the four occurred in the superomedial quadrant. One case occurred inferiorly, rare site for orbital typical dermoids and epidermoids. The fifth case, also inferior, was an example of a cyst within choristomatous lacrimal tissue [7].

Another case of orbital dermoid in an adult female presenting as a monocular elevation deficiency with unilateral ptosis of right eye was reported [8]. It was investigated and managed with good results.

In our case series, four cases with atypical dermoids presentation were reported. Out of the four cases, three were confined to the orbit while one presented as a conjunctival mass. In the orbit, one mass was found over the brow over the frontal bone while the other occurred inferomedially and medially, all unusual sites for dermoid.

A review of 160 CT studies of orbital dermoids revealed that 65% were lateral and 30% were medial to the globe, only one was entirely behind the globe, 85% had changes in adjacent bone, 73% had a visible wall, 27% had a CT attenuation similar to orbital fat, 14% had calcification, 5% had a fluid level, and 20% had abnormal soft tissue outside the cyst [4].

Although in routine practice we usually see typical dermoids in superomedial location or superolateral location. Occurrence of atypical orbital dermoids cannot be ignored. These atypical dermoids although rare do occur. An appropriate surgical approach and histopathological examination can confirm its diagnosis simultaneously managing it adequately.

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